The Rhodoid Nevus: A Proposed Term for a So Far Unnamed Capillary Malformation

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Key Words

Rhodoid nevus • Capillary malformation • Capillary malformation-arteriovenous malformation • *RASA1* mutations • Nevus flammeus

Introduction

In 2003, a distinct autosomal dominant trait characterized by multiple, rather small capillary malformations of round or oval shape was described by Miikka Vakkula's team from Brussels [1]. They found that this phenotype that had previously been mapped to chromosome 5q [2, 3], was caused by *RASA1* mutations. Because the skin lesions were sometimes associated with a fast-flow arteriovenous malformation of the Parkes Weber type, they chose the name 'capillary malformation-arteriovenous malformation (CM-AVM)' [1]. In the meantime this peculiar vascular disorder has become a well-established entity with its own OMIM entry [4].

For clinical dermatologists, however, there still remains a major problem. How can we recognize the rather typical vascular lesions of this phenotype if they have no specific name? So far, all experts are describing the cutaneous hallmark of this autosomal dominant trait as 'capillary malformation' [1–7]. This term is incredibly broad and includes many vascular disorders such as nevus flammeus, nevus anemicus, angiokeratoma circumscriptum as well as nuchal or glabellar salmon patches [8]. As a consequence, the cutaneous lesions of CM-AVM have erroneously been called 'nevi flammei' [4] or 'port-wine stains' [1–4, 6].

For obvious reasons, the capillary malformation associated with CM-AVM syndrome should be identifiable by a separate name. Because the lesion is typically of a 'pink to red' color [1, 5, 7], I propose the designation 'rhodoid nevus'. In ancient Greek, ῥοδοειδής (*rhodoides*) means 'rose-like' or 'rose-colored'. Accordingly, CM-AVM could also be called 'rhodoid nevus syndrome'.

The distinguishing criteria of rhodoid nevus (fig. 1) are summarized in table 1. Its color is lighter than that of nevus flammeus (fig. 2). On the other hand, its hue is usually darker than that observed in nevus roseus (fig. 3), a congenital skin lesion that is sometimes found to be associated with a macular nevus spilus, resulting in phacomatosis spilosea [9–11]. Rhodoid nevi tend to be rather small and of round or oval shape [6, 7], and they show a haphazard distribution [5, 7]. By contrast, both nevus flammeus and nevus roseus are archetypically arranged in a checkerboard pattern [9]. As another distinguishing feature, the rhodoid nevus is often surrounded by a nar-

Fig. 1. Rhodoid nevus in a 12-year-old girl with CM-AVM. A *RASA1* mutation was found in this patient by Dr. M. Vikkula (Brussels, Belgium). Courtesy of Dr. Aïcha Salhi, Algiers, Algeria.

Fig. 3. Distinguishing criteria of nevus roseus are its pale pink color and flag-like arrangement.

Fig. 2. Distinguishing criteria of nevus flammeus are dark red color and flag-like arrangement.

row anemic halo [5, 7]. Moreover, rhodoid nevi are the hallmark of an autosomal dominant trait, whereas both nevus flammeus and nevus roseus usually occur sporadically.

In practice, we should bear in mind that the clinical differences are sometimes not as clear-cut as documented in figures 1–3. For example, nevus flammeus may be rather light-red during infancy and thus be confused with the other two types of vascular nevi. So far, it is unclear whether these vascular lesions can be distinguished by histopathological criteria. In my view, this is rather unlikely. Notwithstanding, the proposed classification appears to be valid.

It should be noted, however, that the list of vascular nevi as presented in table 1 is not complete. Additional examples include cutis marmorata telangiectatica congenita, angiokeratoma circumscriptum and nevus anemicus [8, 12]. Moreover, a peculiar median type of nevus flammeus is found in some patients with Van Lohuizen syndrome (cutis marmorata telangiectatica congenita syndrome) [13]. On the other hand, it should be borne in mind that the fashionable new term ‘capillary malformation’ [1–3, 6, 7, 14] includes several vascular lesions that do not represent nevi, such as the salmon patch and the telangiectatic lesions of Rendu-Osler disease [8].

In conclusion, we should no longer obey the presently prevailing linguistic prescriptivism that conflates dif-

Table 1. Distinguishing criteria of 3 types of vascular nevi

Type of vascular nevus	Color of nevus	Archetypical pattern of nevus	Evolution with time	Syndromic associations	Formal genetic classification	Molecular basis
Rhoid nevus	light red ('pink to red')	haphazard distribution of rather small macules ¹	stable (?)	CM-AVM (= rhoid nevus syndrome)	autosomal dominant inheritance	<i>RASA1</i> mutations
Nevus flammeus	dark red	segmental, flag-like, sometimes systematized	vascular nodules may develop with age	Sturge-Weber-Klippel-Trenaunay syndrome ² ; Proteus syndrome; phacomatosis cesioflammea	usually sporadic; paradominant inheritance	unknown
Nevus roseus	pale pink	segmental, flag-like, sometimes systematized	stable (?)	phacomatosis spilorosea	usually sporadic; paradominant inheritance (?)	unknown

¹ Rarely, a type 2 segmental manifestation in the form of a segmental, flag-like pattern and arteriovenous fistulas of deep vessels is noted in this autosomal dominant trait.

² Includes some cases of spinal involvement (Cobb syndrome) and arteriovenous fistulas (Parkes Weber syndrome).

ferent vascular nevi by simply applying the umbrella term 'capillary malformation'. The proposed new name 'rhoid nevus' will help clinicians to recognize 'CM-AVM', a peculiar vascular phenotype that is inherited as an autosomal dominant trait and caused by *RASA1* mutations [1].

Disclosure Statement

The author has no conflict of interest.

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